A42.E400
JACC March 9, 2010
Volume 55, issue 10A

CONGENITAL CARDIOLOGY SOLUTIONS
(PEDIATRIC CARDIOLOGY AND ADULT CONGENITAL HEART DISEASE)

CLINICAL STUDY ON MORTALITY AND MORBIDITY OF PATIENTS WITH EISENMENGER SYNDROME -JAPANESE MULTICENTER STUDY

ACC Poster Contributions
Georgia World Congress Center, Hall B5
Sunday, March 14, 2010, 3:30 p.m.-4:30 p.m.

Session Title: Adult Congenital Heart Disease
Abstract Category: Adult Congenital Heart Disease
Presentation Number: 1119-404

Authors: *Hisanori Sakazaki, Koichiro Niwa, Makoto Nakazawa, Tsutomu Saji, Toshio Nakanishi, Motoki Takamuro, Michihiko Ueno, Hitoshi Katou, Shinichi Takatsuki, Seiki Matsushima, Namiko Kojima, Fukiko Ichida, Shigetoyo Kogaki, Sachiko Kido, Yoshio Aragaki, Kenji Waki, Teiji Akagi, Kunitaka Jo, Atsushi Souuchi, Kenji Suda, Study committee of Japanese Society of Pediatric Cardiology and Cardiology Surgery, Hyogo prefectural Amagasaki Hospital, Amagasaki, Japan*

Background: Recent success of disease-specific therapy is expected to improve prognosis of patients with Eisenmenger syndrome(ES), however, sudden death(SD) is a major cause of death in ES. Our aim is to clarify predictive factors for sudden death in these patients in current era.

Methods: Data regarding patient characteristic, function class, percutaneous Oxygen saturation (SpO2), electrocardiographic, echocardiographic, laboratory parameters, arrhythmia and systemic complications and medication were collected in 104 patients (male 46,16-66 yrs) with ES who visited outpatient clinic in 14 participated institutes from 1998 to 2009.Clinical data between SD group and non SD group were compared, and prognostic factors for SD were analyzed.

Results: During a median follow-up of 15 yrs, sixteen patients died; SD in 8 (50%) and heart failure in 3 (19%). Causes of SD were unknown (5), ventricular fibrillation(2)and pulmonary haemorrhage(1). In SD group, mean value of SPO2 was lower (76% vs 85%, p=0.001), arrhythmia was more frequent than non SD group (50% vs 19%, p=0.07). However, there were no differences regarding number of NYHA functional class>2 (62% vs 35%,p=0.14), hemoptysis (12% vs 19%,p=0.6), taking bosentan and/or sildenafil (13% vs 29%,p=0.27) In multivariate analysis, SpO2<80% was only factor independently related to sudden death (RR 7.3, p=0.005).

Conclusions: Even in current era of disease-specific therapy, sudden death was still a main cause of death in Eisenmenger syndrome. To prevent sudden death, meticulous care and close follow-up for hypoxia and arrhythmia is necessary.